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Doubly Committed Subarterial Ventricular Septal Defects: Echocardiographic Features and Surgical Implications

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Doubly committed subarterial (supracristal, subpulmonary) ventricular septal defects are often complicated by aortic regurgitation resulting from aortic valve herniation into the defect. The clinical, echocardiographic and catheterization findings in 48 patients aged 0.3 to 46.4 years (median 9.5) with a doubly committed subarterial ventricular septal defect were reviewed. Aortic valve herniation was present in 38 (79%) and 55% of these had aortic regurgitation. The prevalence of both findings increased gradually with advancing age.

The defect was closed surgically in 41 patients. Surgery during the first 2 years of life (median 0.4 year) was performed in 13 patients (group I), mainly because of a large shunt with a pulmonary to systemic flow ratio (Qp/Qs) 3.8 ± 1.4 (mean ± SD). Aortic regurgitation was present preoperatively in two patients (15%), persisted postoperatively in one patient and did not develop in any after repair (median duration of follow-up 2.3 years, range 0.1 to 7.4).

In the other 28 patients (group II) surgery was performed between 4.8 and 46.4 years of age (median 11.5). These patients were generally less symptomatic and had a smaller shunt (Qp/Qs 1.5 ± 0.5, $p < 0.001$). Preoperative aortic regurgitation was present in 18 (64%). It persisted in 15 postoperatively, but in 13 of these it had diminished.

Two-dimensional echocardiography in multiple views identified the site of the ventricular septal defect in all patients. Serial echocardiographic examinations demonstrated the progressive nature of aortic valve herniation, the partial occlusion of the defect by the herniated sinus and the development of aortic regurgitation. These findings suggest that timely surgical closure of these defects may prevent aortic regurgitation.

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Aortic regurgitation is found in 5 to 7% of children with a ventricular septal defect of all types (1–3). It is an important complication because it is often progressive and may be a cause of late morbidity and mortality (2–4). Aortic regurgitation seldom occurs with perimembranous ventricular septal defects, but is common in doubly committed subarterial ventricular septal defects (2,3). The latter are situated adjacent to the pulmonary and aortic valves (5) and are also known as supracristal, subarterial infundibular or subpulmonary or subpulmonic ventricular septal defects. The predisposition of these ventricular septal defects to the development of aortic regurgitation has been attributed to the deficient muscular support of the aortic valve apparatus. During diastole, the bulging adjacent sinus of Valsalva protrudes through the defect (6–8), whereas in systole the left to right shunt carries a portion of the sinus into the defect (8,9). The protrusion of the aortic sinus has been referred to as aortic valve prolapse. Because it is through an abnormal orifice, the term herniation is more precise and will be used throughout this communication.

Many reports (1–4,6–16) have addressed the relation between ventricular septal defects with aortic regurgitation and their clinical features, diagnostic criteria and therapeutic approaches. Only some of these reports (7,9,10,13), however, have focused on doubly committed subarterial defects and their particular predilection for developing aortic regurgitation. The largest study was of Japanese patients (17) in whom a doubly committed subarterial defect accounts for ≤30% of all ventricular septal defects (18). The high incidence of aortic valve involvement in these patients has prompted recommendations for close clinical observation for aortic regurgitation (1,4,6,10,17), repeated aortography

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(13) and early surgical closure of these defects (2,8,15,17). More recently, two-dimensional echocardiography and Doppler ultrasound have been used to define the specific site of a doubly committed subarterial ventricular septal defect as well as the presence of aortic valve herniation and aortic regurgitation (19–23). By virtue of their noninvasive nature, these techniques seem to be ideally suited to follow these patients serially.

Using the various modalities of cardiac ultrasound as well as data from catheterization and surgical observation, the present study was undertaken to determine 1) the prevalence at different ages of aortic valve herniation in children with a doubly committed subarterial ventricular septal defect; 2) the relation of herniation to the development of aortic regurgitation; 3) the effect of early surgical repair on the development or course of aortic regurgitation; and 4) the impact of echocardiographic and Doppler observations on diagnosis and timing of surgical intervention.

Methods

Study patients. From January 1980 through July 1987, 48 patients with a doubly committed subarterial ventricular septal defect underwent diagnostic evaluation at the University of California, San Francisco. Patients whose defect was part of a complex cardiac lesion such as Taussig-Bing anomaly as well as patients with a history of bacterial endocarditis were excluded from the study. Of the 48 patients, 34 were male and 14 female; the median age was 9.5 years (range 0.3 to 46.4). Two patients were seen only once. The median span of observation of the remaining 46 patients was 8.5 years (range 0.3 to 46.4).

We reviewed medical records, echocardiograms, catheterization data, angiograms and surgical reports to document each patient's course and surgical outcome. The presence of aortic valve herniation or regurgitation was noted at the time it was found by echocardiography, angiography or both. The absence of aortic valve herniation was defined accordingly; the absence of aortic regurgitation was based on echocardiographic or angiographic observations in 27 patients and on auscultation in 4.

To determine the onset and course of herniation and regurgitation, we grouped the patients by age at each of their visits and determined the prevalence of aortic valve herniation and aortic regurgitation within each age group. Of the 46 patients followed up serially, 28 had serial echocardiographic and angiographic observations over a period of at least 5 years before surgical intervention. In this cohort followed up longitudinally, we noted the age at which aortic valve herniation and aortic regurgitation were first documented in each patient.

To analyze the influence of early surgery on the development of aortic regurgitation, we placed the 41 patients who underwent operative repair of their ventricular septal defect into two groups according to the timing of their surgery. The first group consisted of patients who required surgery within the first 2 years of life because of a large ventricular shunt with congestive heart failure or severe right ventricular outflow tract obstruction. The second group consisted of patients who underwent operation later in life because they had a smaller shunt and were less symptomatic in infancy and early childhood.

Echocardiography. All patients had at least one echocardiographic examination with the use of Advanced Technology Laboratories Mark 500, 600 or Ultramark 8 scanners, as well as Irex-Aloka 880 or Hewlett-Packard 77020A instruments for Doppler color flow mapping. Two-dimensional imaging and real-time directed M-mode echocardiography were performed with the use of standard parasternal, apical and subcostal transducer locations (19–21). In addition, pulsed Doppler ultrasound was performed since 1982, continuous wave Doppler ultrasound since 1985 and Doppler color flow mapping since 1986. All studies were recorded on 1/2 in. (1.27 cm) videocassette tapes. Cardiac catheterization, including retrograde aortography, was performed in 43 of the 48 patients. In five patients the diagnosis was based solely on echocardiographic findings. The anatomic diagnosis was confirmed by direct observation in all 41 patients undergoing surgical repair.

We reviewed the echocardiographic studies to determine in which views a doubly committed subarterial ventricular septal defect was seen best and to define the incidence of additional findings, such as aortic valve herniation, aortic regurgitation, pulmonary valve flutter and right ventricular outflow tract obstruction. We reviewed the angiograms and the catheterization reports for the same features and for the magnitude of shunts.

Statistics. We used Student's t test and chi-square analysis to evaluate natural history data. The kappa statistic, an estimate of concordance (24), was used to compare the echocardiographic and angiographic findings.

Results

Natural history. The clinical data of the 48 patients with a doubly committed subarterial ventricular septal defect are outlined in Table 1. At some time during the study, aortic valve herniation was present in 38 patients (79%); aortic regurgitation was present preoperatively in 21 (55%) of these 38. The prevalence of aortic valve herniation increased with advancing age as did that of aortic regurgitation, but the former was higher at all age groups (Fig. 1). Aortic regurgitation was present in only 5% of children <3 years old, gradually increased during childhood and reached a prevalence of 78% in adults. Right ventricular outflow tract obstruction was found in five patients, two of whom had a severely obstructive double-chambered right ventricle and three had mild infundibular stenosis. None had valvular

Table 1. Clinical Data of 48 Patients With a Doubly Committed Subarterial Ventricular Septal Defect

Patient No.	Age (yr)/ Gender	Span of Observation (yr)	CHF	Qp/Qs	Age at Surgery (yr)	Aortic Valve Herniation		Aortic Regurgitation		
						Preop	Onset (yr)	Preop	Onset (yr)	Postop
Early Surgery (Group I)										
1	3.8M	3.8	Y	3.0	0.3	N		N		N
2	3.8F	0.7	Y	6.0	0.1	N		N		N
3	3.1M	2.8	Y	3.5	0.5	N		N		N
4	3.3M	3.0	Y	3.3	0.3	N		N		N
5	0.3F	0.3	Y	6.0	0.2	N		N		N
6	2.1M	0.3	Y	5.0	0.2	N		N		N
7	2.8M	2.8	N	3.4	1.1	Y	0.7	Y	0.7	N
8	9.9M	9.3	Y	2.7	1.9	Y	1.8	Y	1.8	Y
9	8.8M	6.9	Y	2.3	1.5	Y	1.2	N		N
10	6.1M	6.1	Y	5.4	0.3	N		N		N
11	1.5F	1.2	Y	3.3	0.4	N		N		N
12	5.5M	5.5	Y	2.1	1.5	N		N		N
13	3.1M	3.1	Y	1.3†	1.6	Y	1.3	N		N
Late Surgery (Group II)										
14	8.8M	8.8	N	1.3	8.8	Y	3.4	Y	8.8	N
15	5.3M	5.0	Y	1.9	5.0	Y	4.9	N		N
16	5.8F	5.1	Y	1.2	5.1	Y	5.0	Y	5.0	Y
17	9.1M	9.1	N	1.0	9.1	Y	3.8	N		N
18	15.8M	14.7	N	1.4	7.7	Y	6.4	Y	6.4	Y
19	19.6M	18.8	N	1.1	15.8	Y	15.6	Y	15.6	Y
20	14.0M	13.3	Y	1.4	13.3	Y	13.0	Y	13.0	Y
21	14.4F	13.7	N	1.0	12.4	Y	7.8	N		N
22	10.8M	10.2	N	1.5	10.2	Y	9.8	N		N
23	13.5M	13.4	N	2.3	8.7	Y	8.5	Y	8.6	N
24	13.5M	13.4	N	1.1	12.3	Y	9.4	Y	11.8	N
25	14.7M	13.8	N	1.1	10.6	Y	10.2	N		N
26	6.7F	6.4	N	1.3	6.4	Y	3.3	N		N
27	19.9M	18.9	N	1.5	16.9	Y	5.2	Y	14.8	Y
28	20.0M	19.0	N	1.1	17.9	Y	17.3	N		N
29	16.3F	16.2	N	1.7	16.1	Y	15.9	N		N
30	9.3M	7.6	Y	2.4	5.3	Y	5.3	N		N
31	23.5M	21.8	N	1.0	17.1	Y	15.8	Y	15.8	Y
32	17.1M	15.0	N	2.0	14.9	Y	14.8	Y	14.8	Y
33	46.4M	46.4	N	1.9	46.4	Y	45.9	Y	45.9	Y
34	5.8M	4.9	N	2.3	4.8	Y	4.6	Y	4.6	Y
35	14.4M	14.3	N	1.0	14.3	Y	14.2	Y	14.2	Y
36	8.0F	7.4	Y	1.9	5.0	Y	1.5	Y	4.0	Y
37	36.1M	35.5	N	1.3	35.5	Y	31.5	Y	31.5	Y
38	16.6F	15.8	Y	1.1	14.9	Y	13.8	Y	13.8	Y
39	9.8M	8.5	Y	1.5†	8.2	Y	8.1	Y	8.1	Y
40	10.4F	10.1	Y	2.2	7.8	Y	7.7	N		N
41	29.6F	28.9	N	1.8	28.9	Y	28.7	Y	28.7	Y
No Surgery										
42	2.8F	2.5	N			N		N		
43	11.8M	*	N			Y	11.8	N		
44	24.3M	*	N			Y	23.3	Y	23.3	
45	26.3F	25.4	N			Y	24.7	N		
46	1.9M	1.9	N	1.6		Y	0.2	N		
47	2.5M	2.2	N	1.7		Y	2.2	N		
48	3.5F	3.2	N			Y	2.2	N		

*One visit only; †double chambered right ventricle. CHF = congestive heart failure; F = female; M = male; N = no; Qp/Qs = pulmonary to systemic flow ratio; Y = yes; postop = after surgery; preop = before surgery.

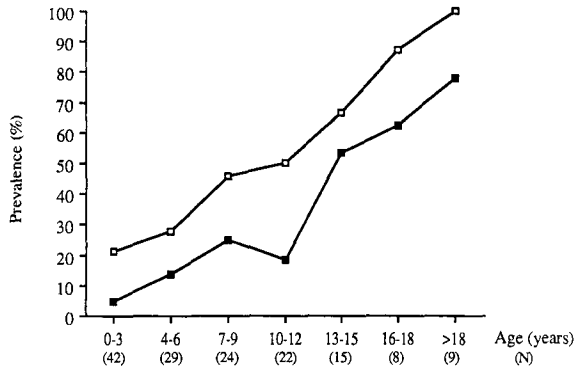

Figure 1. Prevalences of aortic valve herniation (**open squares**) and regurgitation (**solid squares**) proved by echocardiography, angiography or both in 48 patients of various ages with a doubly committed subarterial ventricular septal defect. Prevalences are expressed in percent of the number of patients (N) in each age group.

pulmonary stenosis or subpulmonary obstruction caused by the herniated aortic sinus. All but 2 of the 28 patients, who had been followed up serially over ≥5 years before surgical intervention, were first seen as neonates or young infants and none had aortic regurgitation at that time; 17 of these patients subsequently developed aortic regurgitation (Fig. 2).

Figure 2. Aortic valve herniation and aortic regurgitation assessed by echocardiography, angiography or both in 28 patients with a doubly committed subarterial ventricular septal defect who were followed up for at least 5 years before any surgical intervention. **Horizontal lines** indicate the span of clinical observation for each patient. The symbols indicate the ages at which aortic valve herniation alone (**open circles**) or herniation with aortic regurgitation (**closed circles**) was first documented.

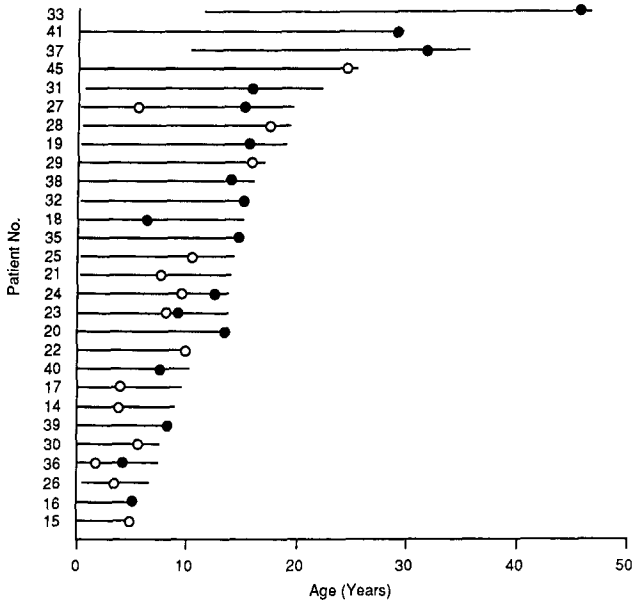

Table 2. Postoperative Aortic Regurgitation in Relation to Preoperative Findings in 41 Patients

	Preop: No AVH, no AR	Postop: AR	Preop: AVH, no AR	Postop: AR	Preop: AVH and AR	Postop: AR
Group I (n = 13)	9	0	2	0	2	1
Group II (n = 28)	0	—	10	0	18	15

AR = aortic regurgitation; AVH = aortic valve herniation; Postop = after surgery; Preop = before surgery. Group I and Group II comprise patients who underwent early and late surgery, respectively.

Early surgical repair before 2 years of age (median 0.4 year) was performed in 13 children (group I). In 12, the primary indication was congestive heart failure due to a large left to right shunt with a pulmonary to systemic flow ratio (Qp/Qs) of 3.8 ± 1.4 (SD) and pulmonary hypertension; in 1, the indication was severe right ventricular outflow tract obstruction (Patient 13).

The remaining 35 patients did not require early surgery. Of these, 28 patients (group II) underwent surgical repair between 4.8 and 46.4 years of age (median 11.5) because of the presence of aortic regurgitation or concern about its development. Their Qp/Qs ratio at the time of surgery was 1.5 ± 0.5 ($p < 0.001$ compared with group I). In group II patients, the surgeon commented on the size of the defect in 25 operative reports. In 22 (88%), the defect was described as being large (that is, ≥1 cm in diameter) but partially or almost completely occluded by the protruding right aortic sinus. Surgery was performed by patch closure of the ventricular septal defect in all but one patient whose small defect was closed by direct suture (Patient 40). Two group II patients (Patients 33 and 35) required plication of the right aortic sinus and cusp, and another (Patient 31) underwent aortic valve replacement. There were no early or late postoperative deaths. Operative morbidity was limited to postpericardiotomy syndrome in four patients and wound infection in one. Seven patients aged 1.9 to 26.3 years (median 3.5) have not yet undergone surgical repair; six have herniation and, of these, one has aortic regurgitation.

Of the nine patients in group I who had neither aortic valve herniation nor aortic regurgitation preoperatively, none developed either feature postoperatively (Table 2). Of the two patients in group I who had herniation but not regurgitation before repair, neither developed regurgitation after surgery. Of the two patients in group I who had both herniation and regurgitation before surgery, one had lesser regurgitation and one had no aortic regurgitation after repair. The median period of follow-up in group I patients was 2.3 years (range 0.1 to 7.4). All patients in group II had aortic valve herniation before surgery. Of the 10 group II patients without preoperative aortic regurgitation, none developed regurgitation postoperatively. In the remaining 18 group II

Table 3. Frequency of Visualization of the Ventricular Septal Defect in Different Views in 48 Patients

View	Percent Visualized
Combination of all views	100
Parasternal long axis	85
Parasternal short axis	85
Subcostal sagittal	61
Apical long axis	20
Subcostal coronal	15
Apical four chamber	10

patients, regurgitation was present preoperatively; it persisted postoperatively in 15, but 13 of these patients had lesser regurgitation than that noted preoperatively. The incidence of aortic regurgitation after surgical repair was significantly higher in group II than in group I patients ($p < 0.01$).

Echocardiographic findings. The site of the ventricular septal defect by two-dimensional echocardiography was recognized in all 48 patients studied preoperatively (Table 3). The defect was most frequently displayed in the parasternal long-axis (Fig. 3), short-axis and subcostal sagittal views (Fig. 4). The defect was always seen in close proximity to both the pulmonary and the aortic valve. Their adjacent sinuses were contiguous without any interposed outlet septum (Fig. 3 and 4). Other views including subcostal coronal or apical four chamber views were less reliable in detecting a doubly committed subarterial ventricular septal defect. Aortic valve herniation was identified by two-dimensional echocardiography in 36 of the 48 patients. The herniation of the right aortic sinus into the right ventricular outflow tract was seen best in the parasternal long- (Fig. 3) and short-axis views (Fig. 5). Varying degrees of herniation of the aortic sinus were seen. Particularly in its early stages, herniation was quite subtle and appeared only as a slightly irregular anterior protrusion of the involved sinus (Fig. 6). When herniation was more severe, the elongated right sinus protruded well into the right ventricular outflow tract, often obscuring the defect almost completely throughout the cardiac cycle (Fig. 3, bottom). In these patients, a portion of the leaflet was distorted or also herniated (Fig. 3, middle).

Systolic pulmonary valve flutter was seen in 29 (76%) of 38 patients studied by M-mode echocardiography (Fig. 7). Furthermore, Doppler flow mapping identified a discrete jet from the ventricular septal defect within the right ventricular outflow tract in all 38 patients studied with pulsed Doppler ultrasound. Doppler color-coded flow mapping was performed in 13 patients. The parasternal short-axis view best demonstrated the position and direction of the jet that emerged from the defect in the right ventricular outflow tract close to the pulmonary valve and passed directly into the pulmonary artery (Fig. 8). This pattern was distinctly different from that found in perimembranous ventricular septal defects, where the jet originates close to the tricuspid valve and is directed toward the inflow or central part of the right ventricle.

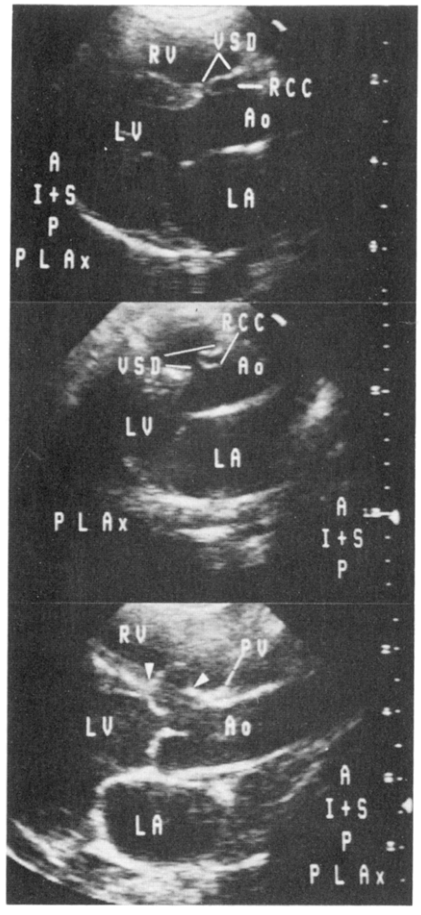

Figure 3. Two-dimensional echocardiographic display of different degrees of aortic valve herniation in three patients with a doubly committed subarterial ventricular septal defect (parasternal long-axis views, PLAx). **Top** (Patient 48), Mild aortic valve herniation with only little anterior deviation of the right sinus of Valsalva is seen. The ventricular septal defect (VSD) is apparently small but is mostly obscured by the herniated sinus and measures actually about 8 mm in diameter. **Middle** (Patient 29), Moderate herniation of the right sinus and the right coronary cusp (RCC) into the ventricular septal defect (VSD) is displayed. **Bottom** (Patient 20), Severe aortic valve herniation into the outflow tract of the right ventricle (RV) is seen in early systole. The **arrowheads** mark the boundaries of the ventricular septal defect, which lies directly inferior to the pulmonary valve (PV) but is completely obscured by the herniated right sinus. A = anterior; Ao = aorta; I = inferior; LA = left atrium; LV = left ventricle; P = posterior; S = superior.

Echocardiographic and angiographic comparison. In 30 patients, aortic root angiography was performed within 1 month of two-dimensional echocardiography. On aortography, aortic valve herniation was seen best in the lateral or left anterior oblique projection as an irregular shape of the involved sinus because of prominence or bulging anteriorly

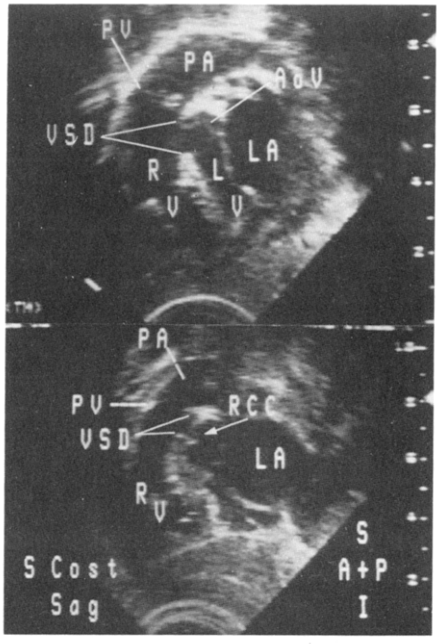

Figure 4. Two-dimensional echocardiographic display of a doubly committed subarterial ventricular septal defect (VSD) in two patients in a subcostal parasagittal view (SCost Sag). **Top** (Patient 11), The pulmonary valve (PV) and the aortic valve (AoV) lie at the same level above the large defect. **Bottom** (Patient 47), In a different patient this defect is covered almost completely by the right sinus of Valsalva. PA = pulmonary artery; other abbreviations as in Figure 3.

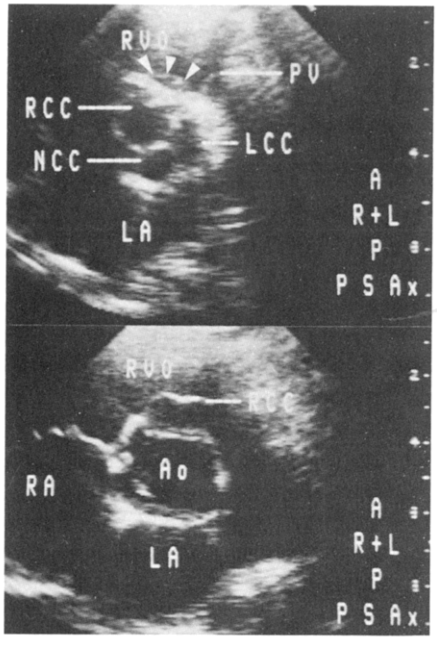

Figure 5. Two-dimensional echocardiographic display of various degrees of aortic valve herniation in two patients with doubly committed subarterial ventricular septal defect. Both frames are shown in the parasternal short-axis (PSAx) view. **Top** (Patient 16), Diastolic frame demonstrating mild aortic valve herniation. The right sinus and coronary cusp (RCC) are enlarged when compared with the left and non-coronary cusps (LCC, NCC). The herniated part of the right sinus (**arrowheads**) is depicted in the right ventricular outflow tract (RVO), close to the pulmonary valve (PV). **Bottom** (Patient 20), Systolic frame showing severe herniation of the right aortic (Ao) sinus. L = left; R = right; RA = right atrium; other abbreviations as in previous figures.

(8,12,13). Aortic valve herniation was seen on echocardiography and aortography in 26 patients, on echocardiography alone in two and on aortography alone in the remaining two; this comparison of two-dimensional echocardiography and angiographic findings of aortic valve herniation shows substantial agreement (κ = 0.660). The herniated sinus was found to be a right sinus in all patients both on two-dimensional echocardiography and on angiography.

Twenty-six of these 30 patients also had a pulsed Doppler ultrasound examination. Aortography and Doppler ultrasound were in agreement for detecting the presence or absence of aortic regurgitation in 22 patients. Doppler ultrasound detected aortic regurgitation in three patients with a negative aortogram, and aortography demonstrated aortic regurgitation in one patient with negative Doppler findings. There was substantial agreement between these methods in detecting aortic regurgitation (κ = 0.655).

Discussion

Natural history. In this study, aortic valve herniation was present in 38 (79%) of the 48 patients with a doubly committed subarterial ventricular septal defect. Its prevalence increased with age, and it was present in all patients >18 years of age (Fig. 1). This increasing prevalence of aortic valve herniation was accompanied by a similar increase in the prevalence of aortic regurgitation, which was present in 55% of the 38 patients and in 78% of the patients >18 years of age. It has been suggested (3,10,12) that aortic regurgitation follows aortic valve herniation by some interval and mainly affects older children or adolescents; however, a large series from Japan (17) based exclusively on angiographic observations showed that both herniation and aortic regurgitation may occur early in childhood. Our study shows similar trends, but because of the noninvasive nature of Doppler echocardiography, we detected herniation even in infancy and regurgitation just after 1 year of age. Our results demonstrate the gradually increasing incidence of aortic valve herniation and regurgitation in a largely non-Asian group of patients. They also affirm that aortic valve herniation precedes the development of aortic regurgitation by an interval that may be very brief (2 months in one of our patients). Although our study might be considered as a series of cross-sectional evaluations, the conclusion regarding de-

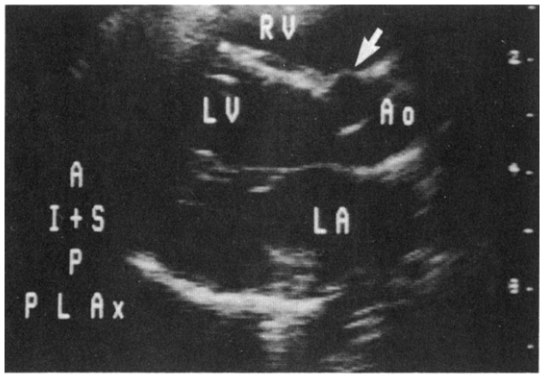

Figure 6. Patient 48. Apparent intact septum in a 3 year old patient with a doubly committed subarterial ventricular septal defect. This parasternal long-axis view (PLAx) shows discrete aortic (Ao) valve herniation. During diastole the ventricular septum seems to be intact. Only a discrete irregularity in the shape of the anterior aortic sinus (**arrow**) indicates its herniation into a ventricular septal defect. Other abbreviations as in previous figures.

velopment of aortic regurgitation is supported by similar findings in the 28 patients followed up longitudinally (Fig. 2).

The term aneurysm of the sinus of Valsalva has been used to describe severe herniation of an aortic sinus in patients with a doubly committed subarterial ventricular septal defect; however, an exact definition as to what size herniation constitutes an aneurysm is somewhat arbitrary. In the report by Momma et al. (17), a sinus of Valsalva aneurysm associated with these defects was notably absent in children but had an increased frequency in adolescents and adults. The finding of an aneurysm in the older and not in the younger patients also suggests that there is an increased severity of herniation with time.

The development of aortic regurgitation may be due to distortion of the herniated sinus by a lack of mechanical barrier at the site of the ventricular septal defect (6–8), or it may result from a long standing Venturi effect through the defect (8,9,17). The involved sinus may also become considerably elongated (Fig. 3). As a consequence of either mechanism, the enlarged protruding aortic sinus may partially obstruct the ventricular septal defect and limit the shunt.

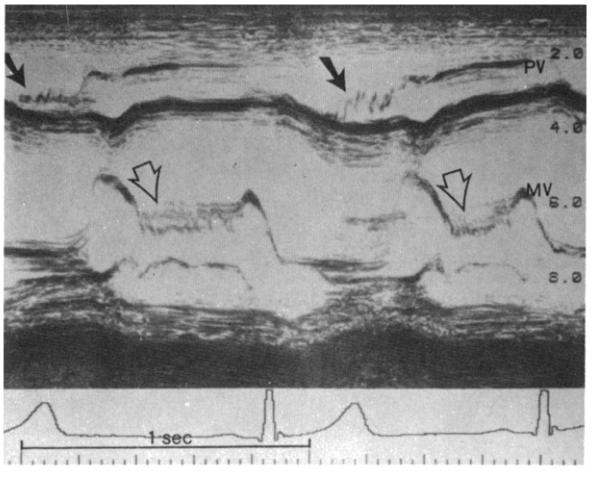

Figure 7. Patient 20. M-mode echocardiogram of the pulmonary valve (PV) and mitral valve (MV) in a patient with a doubly committed subarterial ventricular septal defect and aortic regurgitation. During systole there is flutter of the pulmonary valve (PV) (**black arrow**), which is caused by the systolic jet from the septal defect directed toward the right ventricular outflow tract. During diastole there is flutter of the anterior mitral valve (MV) leaflet (**open arrows**) due to aortic regurgitation.

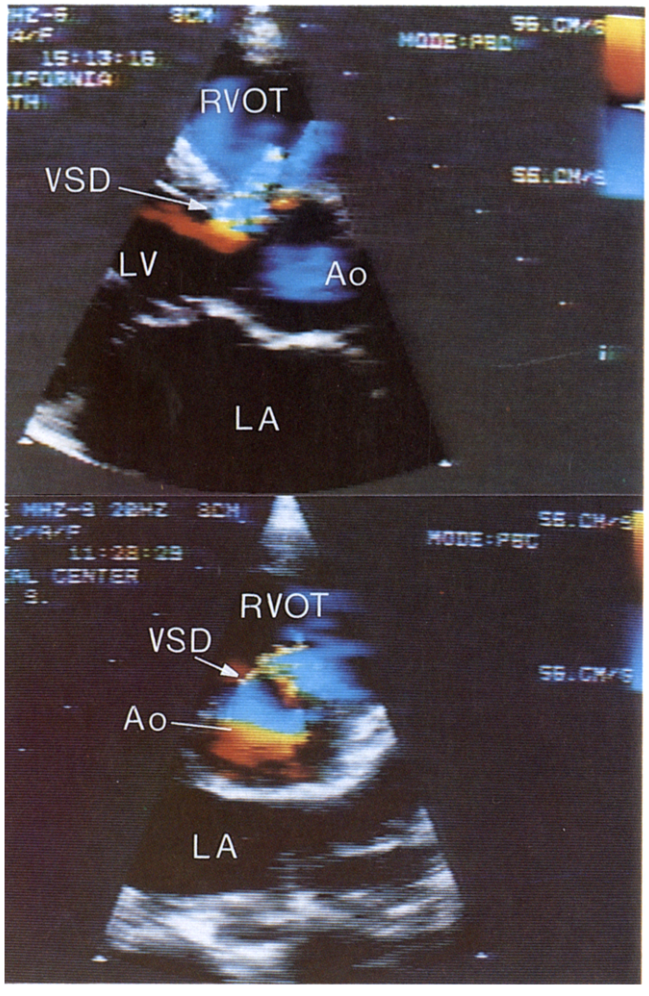

Figure 8. Doppler color flow mapping of the shunt jet in two patients with a doubly committed subarterial ventricular septal defect. **Top** (Patient 29), Parasternal long-axis view showing the shunt flow across the defect (VSD) just below the right coronary sinus directed anteriorly into the right ventricular outflow tract (RVOT). Aliasing of the color signal can be noted because the velocity of the jet increases across the defect. **Bottom** (Patient 46), In a parasternal short-axis view, the jet arises in the right ventricular outflow tract (RVOT) and is directed toward the pulmonary valve. This finding is distinctly different from that in a perimembranous ventricular septal defect. Other abbreviations as in previous figures.

Although our group II patients (those undergoing surgery after 2 years of age) had a small left to right shunt and normal pulmonary artery pressure, most were found at surgery to have a large defect that was partially or almost completely occluded by the right aortic sinus.

Role of surgical repair. If the prevalence of both aortic valve herniation and aortic regurgitation increases with age, one would expect these findings less frequently after early surgical repair. Our data support this supposition. Regardless of the age at repair, none of our patients developed aortic regurgitation postoperatively when herniation without regurgitation was present preoperatively (Table 2), although distortion of the valve mechanism may eventually lead to regurgitation. In contrast, those patients who had both herniation and regurgitation before surgery frequently continued to have aortic regurgitation after repair. The preoperative severity of aortic regurgitation in these patients usually decreased after repair, even though reconstruction of the herniated aortic sinus was performed in only two patients. Therefore, the findings in our surgical series suggest that development or progression of aortic regurgitation may be prevented by early surgical closure of the defect. In contrast, other investigators found no beneficial effect of surgery performed before the onset of aortic regurgitation (3), or recommended deferring surgery to after the age of 5 years and reconstruction of the aortic valve (16).

Limitations. One possible drawback of our natural history study is that it is a retrospective evaluation using different invasive and noninvasive methods to assess aortic valve herniation and regurgitation. This procedure was unavoidable because the period of observation of many patients was long and the diagnostic technology has changed rapidly. Clearly, a prospective study utilizing one technique would be preferable; however, obtaining such information may take decades, and our study currently provides the best information available. Furthermore, given the tendency to surgical intervention, it is unlikely that such a study of an entirely natural history would be possible.

Echocardiographic findings. In the past, angiography has been the principal method of demonstrating doubly committed subarterial defects along with the aortic valve herniation and regurgitation (7,8,13). Documentation of these defects by two-dimensional echocardiography was reported by Sutherland et al. (19) and Capelli et al. (20), while some echocardiographic features of aortic valve herniation were recently reported (22) in a limited number of patients. Our experience with two-dimensional echocardiography in a large number of patients of all ages indicates that the defect and the aortic valve herniation, if present, may be reliably visualized when one uses a variety of views. We found that the ventricular septal defect was imaged best in the parasternal long- and short-axis views (Table 3, Fig. 3 and 5). Other investigators (19) found short-axis scanning to be of only limited value, a difference that may be explained in part by their use of less sensitive ultrasound equipment. The imaging of these defects is quite variable and depends on the degree of herniation. In the absence of herniation, the margins of the defect are usually quite apparent. With minor degrees of herniation, a portion of the defect is obscured, whereas with major degrees the herniated aortic sinus may be immobile and occlude the ventricular septal defect almost completely (Fig. 3, bottom). Rarely, the occlusion may create the impression that the ventricular septum is intact (Fig. 6).

Although the defects often may appear small echocardiographically, they are almost invariably found at surgery to be large and filled by the herniated sinus. For this reason, the size of the defect may be approximated best echocardiographically by measuring the diameter at the base of the herniated portion of the aortic sinus (Fig. 3). Although severe or moderate aortic valve herniation may be readily apparent, great care must be taken to avoid missing minor degrees of herniation (Fig. 6). A discrete irregularity in the shape of the right aortic sinus appearing as a protrusion directed toward the right ventricular outflow tract was a subtle early echocardiographic finding.

Comparison with angiography. In our series, two-dimensional echocardiography proved to be as reliable as aortic root angiography in detecting aortic valve herniation. Aortic root angiography requires special angulation to show this lesion (12,13), and the angle selected for a single aortogram may not be optimal for demonstrating herniation, particularly if it is mild. A tomographic method of imaging with multiple planes such as two-dimensional echocardiography facilitates the demonstration of structures being in close proximity when compared with a projective method such as angiography. In the light of our experience with the echocardiographic findings of aortic valve herniation, we were able to identify, retrospectively, the more subtle angiographic features in many of our patients. Postoperative echocardiograms showed the patch to be acting as a barrier against sinus herniation, even when there was a residual ventricular shunt. The role of the patch as a barrier is probably much more important than complete closure of the defect in preventing aortic valve distortion.

Other echocardiographic features may help establish the diagnosis of a doubly committed subarterial ventricular septal defect, such as the detection of flutter on the pulmonary valve (Fig. 7) (23,25), although this finding may be related to other causes. Doppler flow mapping has proved to be valuable by detecting a flow disturbance directed toward the right ventricular outflow tract in the absence of a obstruction at this site (22). We found Doppler color flow mapping in the parasternal long- and short-axis views to be of considerable value, particularly when the defect is difficult to visualize. It readily depicts the unique location and direction of the shunt jet and greatly facilitates defining a defect almost obscured by a herniated aortic valve (Fig. 8).

Although some investigators (26) have found echocar-

diography to be of limited value for defining herniation in this condition, we found serial echocardiography to be the best method presently available for early detection of aortic valve involvement in these patients. Cardiac catheterization and angiography, if necessary, need only be performed preoperatively.

Timing of surgical repair. Our data indicate that the incidence of aortic herniation and regurgitation increases with age. The increase is gradual, and the risk appears to affect all ages except infancy. Furthermore, progressive herniation of an aortic sinus into a doubly committed subarterial ventricular septal defect precedes the development of aortic regurgitation. Timely closure of this defect decreases the incidence of regurgitation and may obviate the need for aortic valve surgery. In those patients in whom surgery has to be deferred because of patient size or other reasons, serial two-dimensional echocardiography rather than close clinical observation (17) should be the method of choice to detect possible aortic valve involvement.

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